CASE REPORT: BENIGN INTRACRANIAL HYPERTENSION DURING TREATMENT OF DISEASE OF CUSHING (CD) WITH KETOCONAZOLE IN CHILDREN

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Benign intracranial hypertension (BIH) is a syndrome of headache, visual disturbances and papilloedema due to high intracranial pressure with normal brain imaging and composition of cerebrospinal fluid (CSF). BIH have been described after CD control by pituitary surgery or mitotane. We report a case of BIH in a child after Cushing's syndrome control with ketoconazole. Its association to ketoconazole or normalization of cortisol levels with this treatment has not been described yet. Case report: A 12-year-old girl with CD confirmed by elevated urinary free cortisol, cortisol after 1mg dexamethasone suppression of 13.2 µg/dL (n: <1.8), ACTH of 58 pg/mL (n: 10-52), normal pituitary MRI and bilateral simultaneous inferior petrosal sinus sampling with ACTH response to CRH, 26.6 to 250 pg/mL (positive to DC: elevation bigger than 3 times the baseline). Ketoconazole therapy was to control the disease and to obtain aeration of the sphenoid sinus, then be able to undergo transsphenoidal surgery. The doses were gradually incresead based on clinical and laboratory parameters. At the dose of 200mg/day, the patient recovered growth rate, lost 10% of weight and normalized twenty-four hours urinary free cortisol. The patient complained of frontal headache and vomiting after a few days, which was treated as sinusitis (azithromycin for 6 days). No symptomatic relief occurred and a black spot on the right temporal visual field was reported by her. Ophthalmologic evaluation identified bilateral papilloedema. Brain MRI was normal and lumbar puncture confirmed elevated CSF pressure with normal composition. The patient was treated with acetazolamide with resolution of the symptoms and papilloedema. Conclusion: This case emphasizes the association between BIH and ketoconazole therapy in CD.